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Melanotic neuro-ectodermal tumour of infancy

A case report

by

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Summary

A case of melanotic neuro-ectodermal tumour of infancy (retinal anlage tumour, melanotic ameloblastoma) is described. Although a well recognized entity, the biological origin of this tumour is still disputed.

A confusing array of names has been awarded to this tumour which characteristically occurs in infants under one year of age, commonly arising from the maxilla but also from the mandible, frontal bone, mediastinum, shoulder and cerebellum.

While the origin of the tumour remains unclear, the ultra-structural evidence of Haywood *et al.* (1969) supports a neural crest origin. Consistent with this are the findings by Borello and Gorlin (1966) of high urinary vanillylmandelic acid excretion in one case and the correlation of tumour enzyme patterns with those of paragangliomas, neuroblastomas and phaeochromocytomas.

Case Report

A healthy, five-month-old, African male infant was admitted with a swelling arising from the left upper gingival margin. The tumour was first noticed by the mother two months previously and had since grown rapidly. There had been intermittent bleeding from the tumour for a week prior to admission. Local examination revealed a dark, rubbery, firm, lobular tumour, almost black in parts, arising from the anterior three-quarters of the left upper alveolar margin, extending superiorly, laterally and posteriorly into the developing maxilla (Fig. 1).

The tumour protruded out of the mouth but sucking and feeding were only marginally impaired.

Full blood count revealed a haemoglobin of 10.9%, a white cell count of $19 \times 10^9/_L$ (neutrophils 44%, lymphocytes 44%, monocytes 10%, eosinophils 2%).

Radiographs revealed a normal chest. Those of the sinuses showed a large soft-tissue tumour, involving the left side of the face and distorting the antrum, with displacement of teeth and destruction of the alveolar process.

The diagnosis was established by needle biopsy under general anaesthesia, whereupon an examination of the extent of the mass was also undertaken. The core of black tissue obtained was submitted for histological examination and following the diagnosis of melanotic neuro-ectodermal tumour of infancy, definitive surgery was planned.

At surgery there was a thin anterior maxillary wall which was grossly expanded. Complete erosion of the palatine process of the maxilla had occurred, leaving the palatine process of the palatine bone intact. The lateral wall of the maxilla and the lower three-quarters of the posterior wall were totally eroded, the tumour being contained only by the expanded periosteum of the maxilla as it extended into the pterygo-maxillary fissure.

The infraorbital margin of the orbital plate had been eroded but the plate was intact in its posterior two-thirds. The infraorbital nerve and vessels were easily identifiable. The blood supply

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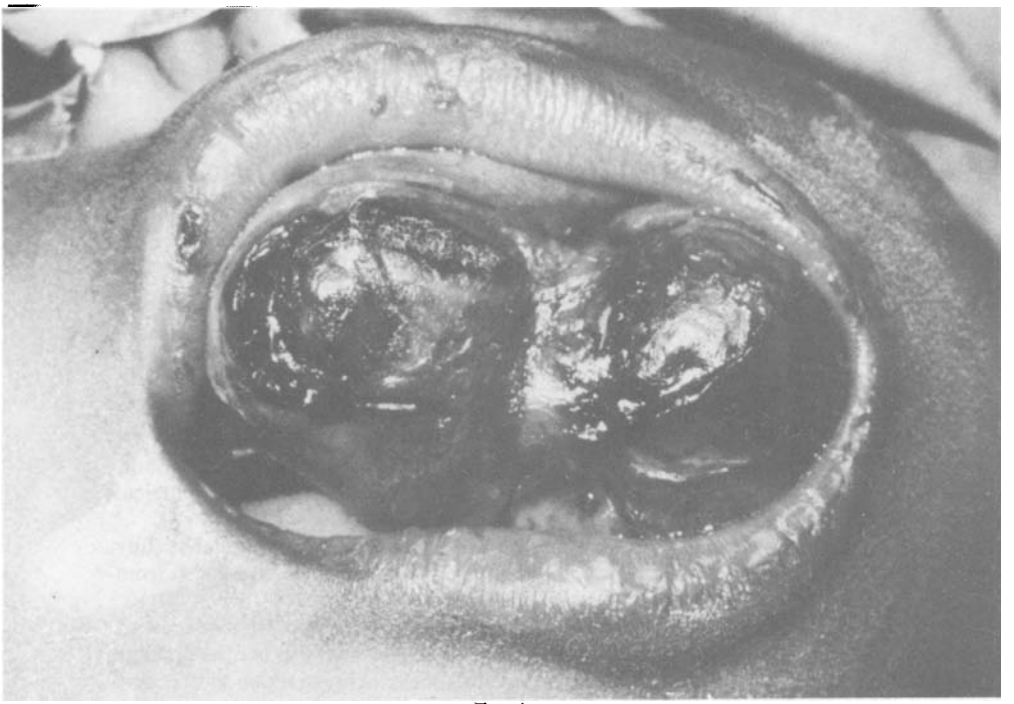


FIG. 1

Photograph of the mouth showing the dark, rubbery black tumour protruding through the lips.

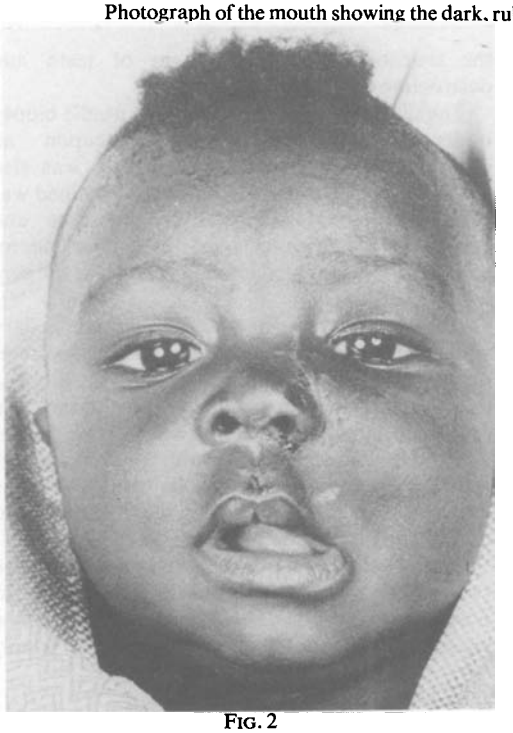


FIG. 2

Photograph of the child three weeks post-operatively.

of the tumour appeared to have arisen from the left greater palatine artery and possibly also from the long sphenopalatine vessel. A left lateral rhinotomy incision was placed, splitting the upper lip in the midline. The soft tissue of the anterior face was elevated and dissected away from the tumour. Once bony and periosteal attachments of the tumour had been divided, it was mobilized with great ease with an index finger from below and behind, and delivered through the incision. It appeared to be encapsulated except for its attachment to the alveolus. The mucosa of the floor and lateral wall of the nose was intact.

Closure in the mouth was effected by interrupted chromic sutures between the greatly expanded buccal mucous membrane and the mucous membrane of the hard palate. Systemic antimicrobials were used and feeding was by gavage for eight days post-operation. Healing was satisfactory and, after three weeks, there was minimal facial deformity and the child was discharged in good health (Fig. 2).

Pathological Findings

The resected specimen (Fig. 3), measured approximately 7×5×4 cm. Much of the tumour appeared to be well encapsulated but one margin

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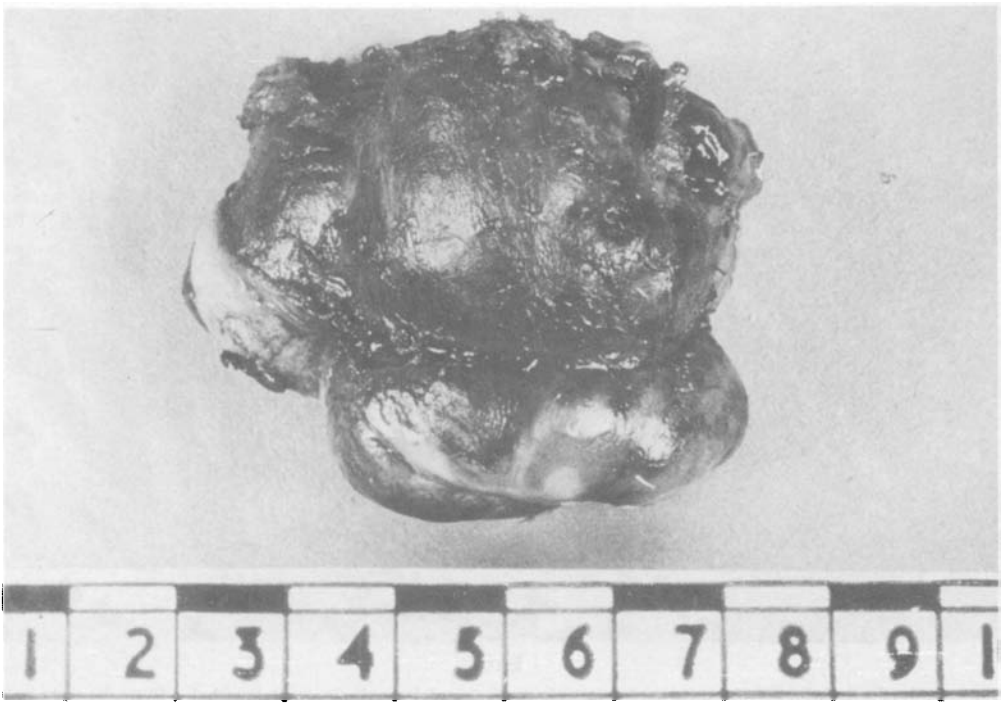


FIG. 3

The resected specimen measuring approximately 7×5×4 cm.

was notably irregular. The cut surface showed lobulation and mottled black pigmentation.

The tumour was of firm consistency, and ill-defined areas of bone were present in the region of the irregular margin. Histological examination confirmed the needle biopsy diagnosis. The cellular element of the tumour was dispersed in a prominent collagenous stroma which had a focal fibrillar appearance (Fig. 4).

Tumour cells were of two types: larger pale-staining polyhedral to cuboidal epithelial-like cells, and smaller neuroblast-like cells with hyperchromatic nuclei and scanty cytoplasm. The epithelial-like cells were arranged predominantly in compact groups and irregular strands but were also seen lining clefts and duct-like spaces. Many of these cells contained abundant coarse intracytoplasmic melanin granules, which was confirmed with the Masson-Fontana silver technique. The neuroblast-like cells occurred as small collections in the stroma, in places surrounded by the polyhedral cells. Bone infiltration was noted at the margin of the tumour.

Discussion

Since the tumour was first described by Krompecher in 1918, more than fifty cases have been reported in the literature under a number of synonyms, including congenital melanocarcinoma, melanotic progonoma, pigmented or melanotic ameloblastoma, pigmented epulis, retinal anlage tumour and melanotic neuroectodermal tissue of infancy. The histological typing of the tumour by Lindborg and Kramer (1971) for the World Health Organization leaves us in no doubt about the characteristics but the histogenesis is still something of an enigma. Theories of origin (Allen *et al.*, 1969) include:

(i) congenital melanoma;
(ii) odontogenic origin;
(iii) retinal anlage origin;
(iv) neuroectodermal development;
(v) keratoma;
(vi) neural crest origin.

Of the above, the possibility of neural crest origin seems the most likely. Borello and Gorlin (1966) put forward some plausible ideas in support of this theory, including the isolation of vanillylmandelic acid in the urine of their case.

Kondstaal *et al.* (1968) studied the histochemical enzyme pattern of the tumours in two patients, comparing it to tumours thought to be

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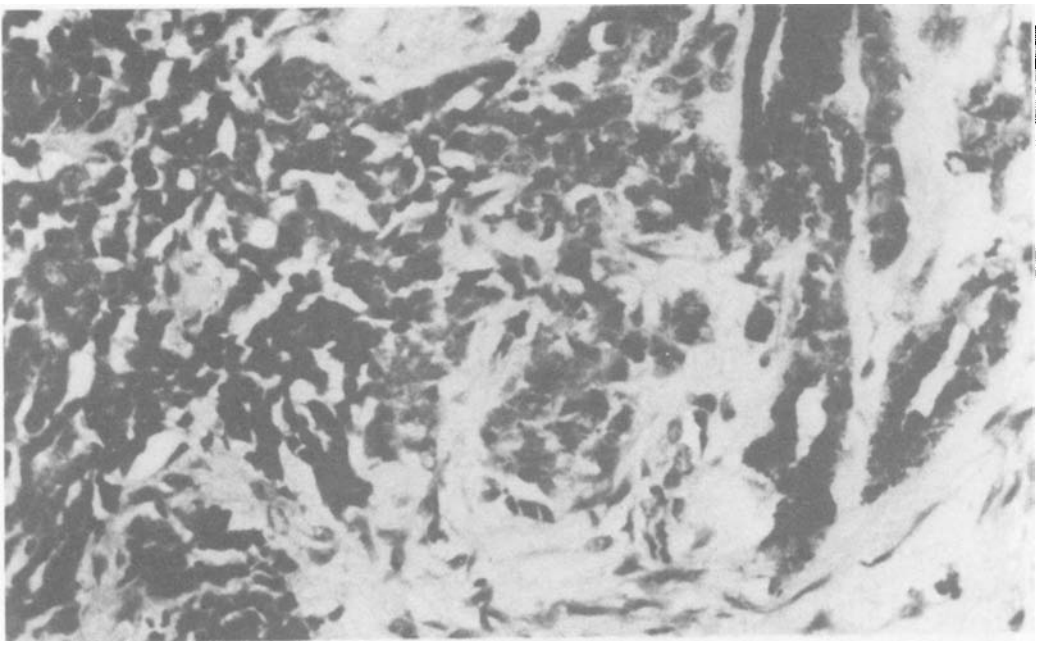


FIG. 4

Photograph of a paraffin section showing larger pale-staining polyhedral to cuboidal epithelial-like cells and smaller neuroblast-like cells with hyperchromatic nuclei and scanty cytoplasm.

derived from the cells of the neural crest, and they found the correlation very close, with the enzyme pattern of the melanotic neuro-ectodermal tumours resembling that of the malignant melanoblastoma, the paragangliomas and phaeochromocytomas, the enzyme pattern of the small cells resembling, to some degree, the cells of the neuroblastomas.

Ultrastructural findings by both Haywood *et al.* (1969) and by Misugi *et al.* (1965) support the hypothesis that the tumour is of neural crest origin.

The tumour occurs in infants, usually under one year of age: the site of origin is frequently the anterior part of the maxilla, other sites being the anterior mandible, anterior fontanelle, shoulder, posterior mediastinum and epididymis; its behaviour is benign. These facts and the fact that recurrences are uncommon and always local, enables one to standardize therapy which, in most hands, is surgery. Simple rather than radical excision suffices (Thoma, 1972), and chemotherapy or radiotherapy are not advised. Further conservative excision may be required for recurrences. Certainly repeated local excision is preferable to primary radical excision which may be mutilating.

The prognosis is good, even after several local recurrences, although dental deformity is always a likelihood.

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